Down's syndrome with lichen nitidus and segmental vitiligo

Sir,

Down's syndrome (trisomy 21) is a multisystem disorder with a birth incidence of approximately 1 in 700 live births, making it the most common autosomal chromosomal disorder causing mental retardation. Down's Syndrome is also associated with an increased incidence of cutaneous disorders.[1-4] In particular, there seems to be an increased incidence of xerosis, atopic dermatitis, seborrheic dermatitis, cutaneous infections, alopecia areata, vitiligo, syringomas, elastosis perforans, keratoderma palmaris et plantaris, and pitryiasis rubra pilaris. We describe a four-year-old girl with Down's syndrome, with segmental vitiligo and lichen nitidus.

A four-year-old female child with Down's syndrome [Figure 1] presented to us with white patches on the right waist and inguinal area for the last one month. She also had asymptomatic papules on the legs for the same duration. She did not have any systemic complaints. On examination, she had segmental vitiligo [Figure 2] on the right waist. The vitiliginous macule had an irregular border and leucotrichia. She also had multiple, discrete, flat, round, smooth, skin-colored-to-slightly pink papules of 1 to 2 mm size, distributed symmetrically over the extensors of both the legs and thighs [Figure 3]. The rest of the skin, hair, and nails were normal. Manifestations of Down's Syndrome in our case were hypertelorism, depressed nose, epicanthal folds, high-arched palate, flat occiput, low-set ears, hypotonia, sandle toe, flat feet, clinodactyly, and delayed milestones. Histopathology of the papules on the leg showed features consistent with lichen nitidus [Figure 4]. It showed orthokeratosis, focal parakeratosis, and thinning of the epidermis just below the parakeratosis. There was no hypergranulosis, focal infiltrate of lymphocytes, histiocytes in the papilla engulfed by the rete ridges, or perivascular lymphocytic infiltration. The rest of the dermis and subcutaneous tissue was normal. Echocardiography, ophthalmological evaluation, and the biochemical and hematological parameters were within normal limits.

There are many reports to show the association of vitiligo with Down's syndrome.[3,4] Our case was associated with segmental vitiligo and lichen nitidus. In literature, there are only two case reports that show the association of lichen nitidus with Down's syndrome.[5,6] Lichen nitidus was generalized in both the cases; one of these cases[6] was also associated with megacolon. Lichen nitidus,[7] a disorder of unknown etiology, is composed of multiple, discrete, smooth, flat, round, flesh-colored to slightly-pink papules. The lesions may occur anywhere over the skin; however, the most frequent sites of predilection are the flexural surfaces of the arms and wrists, lower abdomen, breast, the glans, and shaft of the penis. However, in our case, the lesions were confined to the legs and thighs only. Lichen nitidus is usually asymptomatic; however, pruritus is occasionally present. The histopathology of lichen nitidus is usually distinctive. Keratosis pilaris, lichen spinulosa, lichen scrofulosorum, and verruca

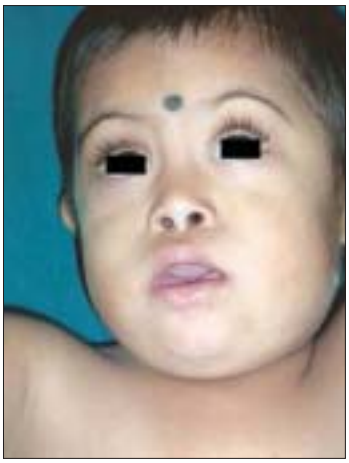

Figure 1: Facies of the child with Down's Syndrome

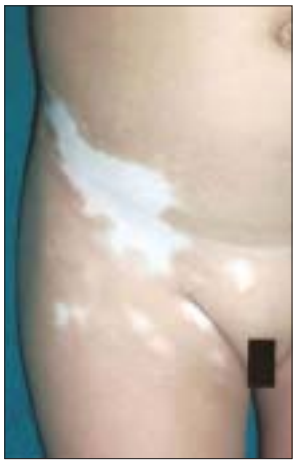

Figure 2: Depigmented macules (segmental vitiligo) on the right waist and inguinal area

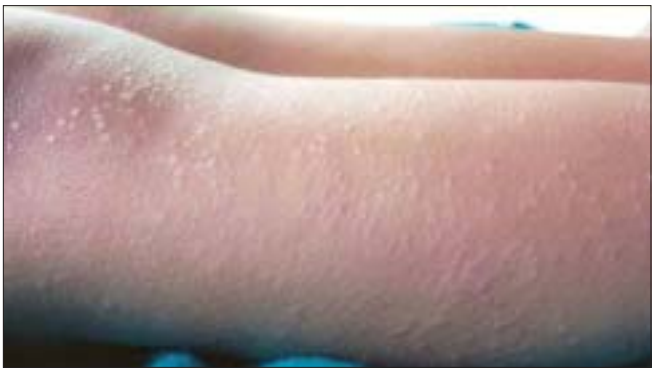

Figure 3: Discrete papules on the legs and thighs

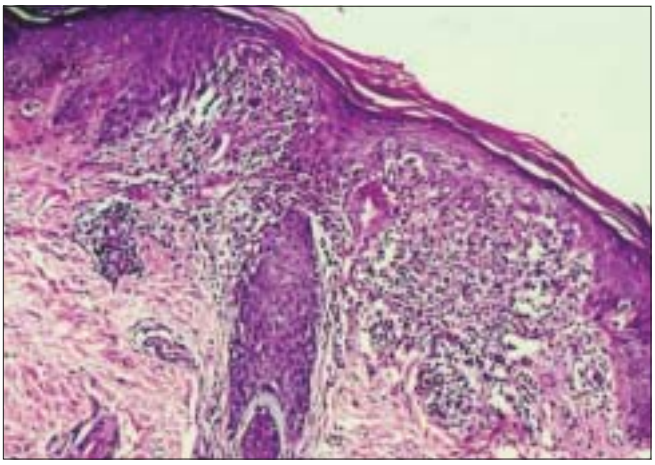

Figure 4: Circumscribed lymphocytic and histiocytic infiltrate in the papillary dermis engulfed by rete ridges situated directly beneath the thinned epidermis with central parakeratosis, loss of granular layer, (H and E, x10)

plana can be considered in the differential diagnosis of lichen nitidus. Keratosis pilaris and lichen spinulosa usually exhibit more keratotic and scaling lesions, unlike the smoother surfaced papules of lichen nitidus. Verruca plana are more variable in size, have a more verrucous surface, and are less likely to involve multiple sites. The lesions in lichen scrofulosorum are perifollicular, grouped, keratotic papules, which occur in young patients with tuberculosis.

Our patient was unique, in that she showed the association of segmental vitiligo and lichen nitidus with Down's syndrome. While it is not unusual to encounter Down's syndrome with vitiligo, to the best of our knowledge, this is the third case of lichen nitidus seen in the patient of Down's syndrome. However, such a combination is not surprising in view of the well-known tendency of the skin of individuals with Down's syndrome to be associated with other dermatological disorders.

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DOI: 10.4103/0378-6323.57738 **- PMID:** 19915257

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